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Non-motor symptoms in childhood dystonia? New perspectives

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Dystonia is a movement disorder characterized by unwanted muscle spasms resulting in involuntary movements and abnormal postures, and is mostly unremitting.¹ Childhood dystonia often spreads from a limb to the trunk with at least two other body parts also included (i.e. it is generalized). This contrasts with adult dystonia which typically is focal or spreads only to adjacent muscle groups. Dystonia is traditionally divided into primary (in which dystonia is a primary feature and is caused by a specific genetic mutation) or secondary, where dystonia occurs secondary to a lesion to the motor system. This could be because of neurological disease, head injury, viral/bacterial infections, and environmental factors causing to insult to the brain, particularly affecting the basal ganglia. The basal ganglia's connections to the prefrontal cortex point to potential deficits in complex cognitive skills and behavioural regulation. Dystonia in childhood can be severely debilitating, affecting the critical development of motor skills useful for interaction and social participation, as well as impacting brain regions associated with emotional functioning. However, comprehensive investigations of non-motor symptoms in childhood dystonia are scarce.

Coenen et al.'s systematic review² is highly relevant and its conclusions provide evidence of a growing research base in non-motor features of childhood dystonia. The extent of cognitive impairment in childhood dystonia is still relatively unknown in comparison to adult dystonia. However, the review provides some insight into current understanding of cognition in child dystonia and the current state of research in the field. Although the review details findings from studies assessing memory, general intelligence, and social cognition, it emphasizes a general lack of empirical data on cognitive impairment in childhood dystonia. Specifically, the review details studies in children that have reported mostly intact cognitive ability in primary dystonia, with mild deficits in working memory and processing speed, but more pronounced deficits in secondary dystonia. Namely, general cognitive ability, social cognition (theory of mind), perceptual reasoning, and visual and verbal memory. Some studies have shown that cognitive ability (except for perceptual reasoning which improved significantly) remains relatively stable after deep brain stimulation surgery in 5- to 18- year-olds with secondary dystonia.³ Thus, the relationship between motor and non-motor impairment remains unclear. It remains to be seen whether the severity of cognitive impairment increases with the severity of motor impairment in childhood dystonia.

Further examinations of cognitive functioning across childhood would provide a much-needed longitudinal perspective on outcomes of childhood dystonia, particularly regarding how cognitive ability impacts

social and emotional functioning and well-being. It is evident that children with dystonia, particularly secondary dystonia, have some cognitive difficulties, and it is becoming apparent that these cognitive difficulties may impact aspects of social cognition, such as interaction and social communication. Studies have reported some difficulties with mentalizing ability and social functioning;⁴ as well as delayed patterns of theory of mind (particularly first-order false belief) development in dystonic cerebral palsy.⁵ Such findings suggest that some children with dystonia may be at risk of experiencing problems with peer relationships and emotion understanding, which play a role in emotional well-being and quality of life.

Overall, although the studies described in Coenen et al.'s review provide evidence that children with dystonia do present with some cognitive difficulties, other cognitive domains such as attention, executive functioning, and language are yet to be sufficiently assessed in this clinical population. Moreover, as noted by Coenen et al., the lack of consistency in administration of standardized neuropsychological tests in childhood dystonia studies suggest that methodological consensus is needed. This is further emphasized by the lack of appropriate control groups included in such studies. Nevertheless, Coenen et al.'s findings open a dialogue on the implications of cognitive deficits for daily functioning and well-being in this clinical population, with respect to the impact it has on social relationships, quality of life, and academic

performance. They also raise an interesting question of whether non-motor symptoms are central to the phenotype of dystonia.

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